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Answer to Case of the Month #149 Alveolar Soft-Part Sarcoma

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Clinical Presentation

A 39-year-old man presented to the emergency department with a 2-week history of a dry cough that was unresponsive to a course of antibiotics. He also suffered from fatigue and believed he had lost some weight over the past 3 months. He had no other specific symptoms and had no significant past medical or surgical history. He was on no medications and was a nonsmoker. On examination, he was apyrexial and his general physical examination was normal apart from the presence of a soft-tissue mass behind his left knee. A full blood count as well as routine serum biochemical analysis was normal. A chest radiograph was performed that showed multiple lung metastases (Figure 1). A magnetic resonance image (MRI) of the left knee was performed.

Figure 2A is a sagittal T1-weighted MRI image showing a 5 × 7 cm soft-tissue mass lying posterior to the knee. The mass involved the posterior muscle compartment. There was no osseous involvement and the femoral artery and vein were separate from it. An unusual feature of the mass was the presence of multiple large vessels present throughout the mass. Figure 2B is a transverse T1-weighted image again showing the mass involving the posterior muscle compartment. Figure 2C is a transverse gadolinium-enhanced image showing marked enhancement of the mass as well as again showing the presence of multiple vessels within the mass. Figure 2D is a T2-weighted image of the mass. The differential diagnosis at this stage included both a large arteriovenous malformation, as well as an alveolar soft-part sarcoma (ASPS). The presence of multiple lung metastases excluded an arteriovenous malformation. In addition, multiple metastases are a characteristic feature of ASPS and therefore this was believed to be the most likely diagnosis. The patient underwent a biopsy of the mass and it was confirmed to be an ASPS (Figure 3).

Diagnosis

Alveolar soft-part sarcoma.

Discussion

ASPS is a rare musculoskeletal tumour that often remains asymptomatic until late in the disease process. The tumour has some characteristic imaging features that allow for a correct

Figure 1. Posteroanterior chest radiograph shows multiple rounded densities throughout both lungs consistent with multiple metastases.

Key Words: Alveolar; Sarcoma; Vascular.

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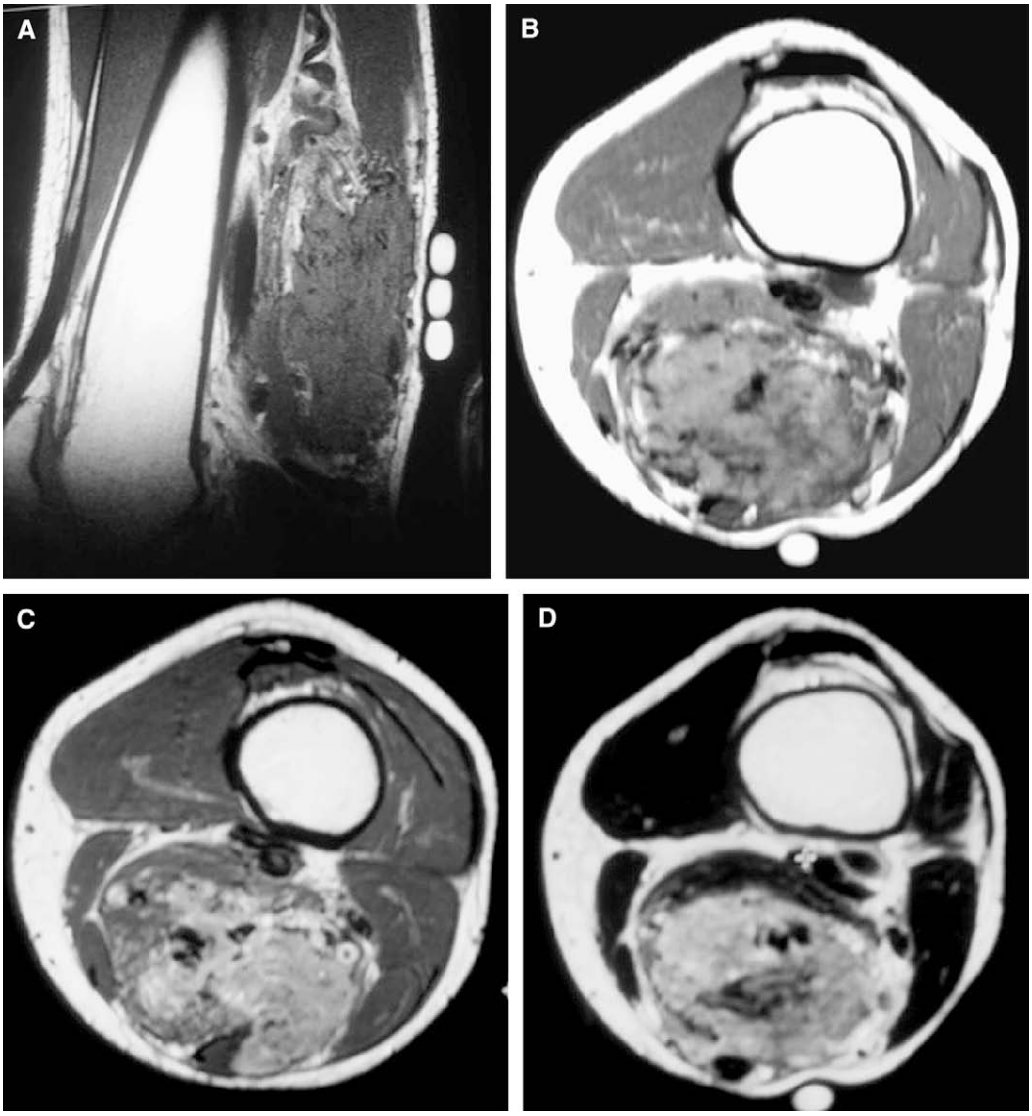

Figure 2. (A) Sagittal T1-weighted image through the left distal femur shows a 5 × 7 cm soft-tissue mass. The mass lies predominantly within the posterior muscle group and on T1 imaging is isointense to muscle. In addition, note the multiple large vessels present throughout the mass. (B) A transverse T1-weighted image again showing the markedly vascular mass, which is isointense to muscle. (C) A transverse gadolinium-enhanced T1-weighted image showing marked enhancement of the mass as well as again showing the presence of multiple vessels within the mass. (D) A transverse T2-weighted image showing the high signal mass posterior to the distal femur.

preoperative diagnosis in many cases. We present a case of a 39-year-old patient who underwent MRI imaging of a soft-tissue mass of the left knee that was believed to represent an ASPS on imaging and was confirmed after core biopsy.

ASPS originally was described by Christopherson [1] and subsequently by Smetana [2]. It is characterized by uniform, organoid nests of polygonal tumour cells, with fibrovascular septa separating them, and delicate capillary-sized vascular channels. There is prominent dyscohesion in these nests, leading to the distinctive pseudoalveolar pattern after which it is named [3].

Cytogenic analysis of ASPS has identified multiple abnormalities that strongly suggest a role for chromosomal abnormalities in the pathogenesis of this tumour [4].

ASPSs are an extremely rare type of tumour and account for fewer than 1% of all sarcomas [5]. These malignancies most often arise in the limbs, head and neck region, or trunk, and occasionally develop as an intracranial mass [6]. They also have been described in the lung [7], stomach [8], breast [9], female genital tract [10], pituitary gland [11], and mediastinum [12]. ASPSs have accounted for approximately 10% of cases reported from large sarcoma referral centers, in both adults [13] and children [14]. Up to 60% of ASPS cases are seen in women [13].

Radiologic investigation is instrumental in reaching a diagnosis of ASPS. Correct diagnosis includes not only the detection but also the characterization and staging of the lesions. Although conventional radiographs may show a soft-tissue mass, ultrasound shows poor specificity and computed tomography shows poor contrast resolution. Conventional angiography may show a hypervascular lesion but MRI, thanks to its high-contrast resolution and multiplanar capabilities,

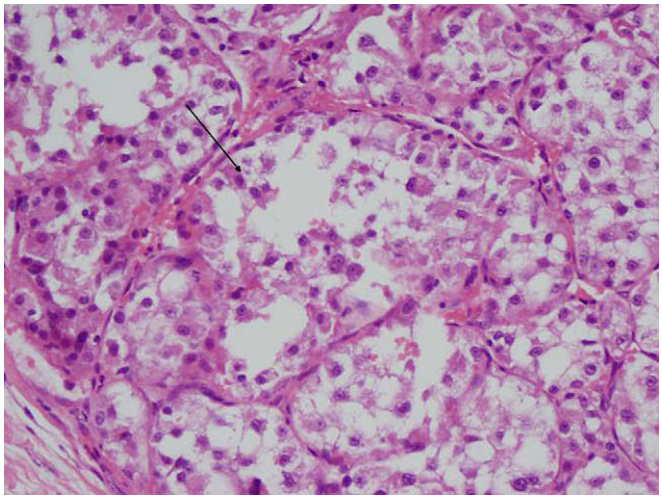

Figure 3. Core biopsy of the mass shows organoid nests of tumour cells with centralcellular dyscohesion resulting in a pseudoalveolar pattern (arrow) (hematoxylin-eosin, 20×).

provides the most information. Iwamoto et al [15] showed that although most soft-tissue sarcomas are isointense relative to muscle on T1-weighted images, 90% of their patients with ASPS showed high signal on both T2- and T1-weighted images. In their group of patients, flow voids were seen centrally and at the margins of the tumours studied [15]. Suh et al [16] also described high signal on T1-weighted images and signal voids but also found that angiographic studies frequently showed enlarged vessels, arteriovenous shunts, and delay of washout. The finding of prominent or enlarged vessels is by no means pathognomonic and arteriovenous malformation, hemangiopericytoma, hemangioendothelioma, synovial sarcoma, rhabdosarcoma, and metastatic deposits among others should be included in the differential diagnosis.

Usually ASPSs are painless, highly vascular, slow-growing tumours. If symptoms are present, they tend to be related to the direct mass effect of the tumour [6]. Often the patient may present with symptoms from metastases, particularly to lung and brain. The primary malignancy rarely is diagnosed before metastases have occurred [17]. The median survival for patients without metastases is 11 years versus 3 years for those with metastases [13].

In the absence of metastatic spread, resection of the primary tumour is the best treatment [18]. In a series by Lieberman et al [13], the median survival of patients who underwent removal of the tumour was 218 months compared with 63.5 months for those who did not. Complete microscopic resection is essential in localized ASPS, but incomplete excision and misdiagnosis frequently occurs [19]. Adjuvant chemotherapy does not seem to be effective in the treatment of ASPSs, although there may be a role for adjuvant radiotherapy in decreasing the risk of local recurrence [13,20,21].

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